Vasculitis
This booklet provides information and answers to your questions about this condition.
What is vasculitis?

Vasculitis means inflammation of the blood vessels. There are several different types of the condition, many with unknown causes, but treatments can be very effective. In this booklet we’ll briefly explain the main types of vasculitis, how it’s diagnosed and treated, what you can do to help yourself and where to get more information.

At the back of this booklet you’ll find a brief glossary of medical words – we’ve underlined these when they’re first used in the booklet.
What’s inside?

2 Vasculitis at a glance
6 What is vasculitis?
8 What are the symptoms of vasculitis?
9 What types of vasculitis are there?
   – Takayasu’s arteritis
   – Giant cell arteritis (temporal arteritis)
   – Polyarteritis nodosa (PAN)
   – Kawasaki disease
   – Wegener’s granulomatosis
   – Churg–Strauss syndrome
   – Microscopic polyangiitis
   – Cryoglobulin-associated vasculitis
   – Henoch–Schönlein purpura
13 Who gets vasculitis?
14 What causes vasculitis?
15 What is the outlook?
15 How is vasculitis diagnosed?
   – What tests are there?
17 What treatments are there for vasculitis?
   – Drugs
19 Self-help and daily living
   – Exercise
   – Diet and nutrition
   – Stop smoking
   – Keep warm
22 Research and new developments
23 Glossary
25 Where can I find out more?
28 We’re here to help
What is vasculitis?
Vasculitis means inflammation of the blood vessels. It can affect any of the body’s blood vessels, causing a variety of different symptoms and problems.

What are the symptoms?
The main symptom of vasculitis is inflammation, and this can be painful. With many forms of the condition the inflammation is internal and you can’t see it. Because vasculitis takes different forms, the symptoms vary from person to person. Many people with vasculitis feel unwell with fever, sweats, fatigue and weight loss. Sometimes these can be the first symptoms you feel, so it’s important to be seen by your GP.

Other symptoms vary according to which part of the body is affected, for example:

- **skin** – rash of spots that can rupture leaving open sores (ulcers)
- **lungs** – coughing or breathlessness
- **nerves** – tingling (pins and needles) or weakness in the arms and legs
- **kidneys** – problems passing urine or blood in the urine
- **fingers and toes** – can turn white or blue, tingle or hurt when exposed to cold conditions.

Headaches, pain in the jaw and problems with the eyes (such as double vision or blurring) are all possible symptoms of a type of vasculitis called giant cell arteritis (GCA).

What types of vasculitis are there?
Vasculitis can be primary, where it occurs on its own, or secondary, where it occurs with another condition. There are several types of primary vasculitis, including:

- **Takayasu’s arteritis** – affects the main artery from the heart (usually in younger women).
- **Giant cell arteritis (temporal arteritis)** – affects the large arteries that supply the head and neck, especially the temporal artery (very rare in people under 50).
• **Polyarteritis nodosa** – inflammation in the medium-sized arteries, such as those supplying the gut and kidneys.

• **Kawasaki disease** – affects small and medium-sized arteries in young children (aged under 5 years).

• **Wegener’s granulomatosis** – involves inflammation of the skin, lungs, eyes and kidneys and swellings called granulomata.

• **Churg–Strauss syndrome** – asthma developing in adult life, followed by inflammation of the blood vessels and swellings called granulomas; this form of vasculitis usually affects the nerves, causing weakness, pins and needles and numbness.

• **Microscopic polyangiitis** – causes kidney problems that can lead to kidney failure, and can also involve the lungs, with bleeding that can cause breathlessness.

• **Cryoglobulin-associated vasculitis** – causes a reduction in blood flow or even a blockage, causing damage to the organs or body tissues.

• **Henoch–Schönlein purpura** – affects the small blood vessels (capillaries) and mostly affects children between 2–10 years; symptoms include a skin rash, arthritis, abdominal pain and/or vomiting or passing blood in the stools, passing blood in urine, fever, headaches and loss of appetite.

**How is it diagnosed?**

Vasculitis is diagnosed using blood tests, x-rays or other scans and biopsies (obtaining a tissue sample to look at under a microscope).

**What tests are there?**

The following tests may be used to check for types of vasculitis:

• blood tests – to test for inflammation, low blood count, kidney and liver function, and immunological blood tests to test for antineutrophil cytoplasmic antibodies (ANCA)

• urine tests – to test for kidney inflammation
x-rays, computerised tomography (CT) and magnetic resonance imaging (MRI) scans – to test for chest or sinus problems

echocardiogram and electrocardiogram (ECG) – to check your heart is healthy

biopsy – to check if your temporal arteries, liver, kidneys, muscles or lungs are affected

ear, nose and throat (ENT) examination – to check if these parts of the body are affected

angiogram – to see which blood vessels are involved.

What treatments are there?

Treatments for the various types of vasculitis include:

- drug treatments, including corticosteroid tablets or injections, immunosuppressive drugs, immunoglobulin injections, antiviral treatments and antibacterial drugs
- plasma exchange (sometimes called blood washing), which may be needed by a small minority of people with the most severe forms of vasculitis
- dialysis in cases of severe kidney damage.

What else might help?

There are some things you can do to help yourself if you have vasculitis, including:

- stopping smoking
- balancing regular exercise with rest
- eating a healthy balanced diet, including enough calcium and vitamin D if you’re on steroids
- keeping warm (especially if you experience Raynaud’s phenomenon)
- understanding the nature of the treatment you’re on and the precautions that you need to take, including immunisations against seasonal flu and pneumonia.
What is vasculitis?
Vasculitis means inflammation of the blood vessels. It can affect any of the body’s blood vessels, causing a variety of symptoms and potential complications.

Blood vessels are the tubes that carry blood around your body. There are three types of blood vessel which can be affected by vasculitis (see Figure 1):

**Figure 1: The three types of blood vessel that can be affected by vasculitis**

<table>
<thead>
<tr>
<th>Arteries</th>
<th>Veins</th>
<th>Capillaries</th>
</tr>
</thead>
<tbody>
<tr>
<td>take blood from the heart to various parts of the body such as organs (e.g. kidneys) and body tissues (e.g. skin)</td>
<td>take blood back to the heart</td>
<td>tiny vessels between the arteries and the veins where oxygen and other materials pass from the blood into the tissues</td>
</tr>
</tbody>
</table>

**Figure 2 The blood circulatory system**

The small vessels, which are too small to be seen with your eye, are spread throughout the tissues in your body.
The organs and tissues in your body need a regular blood supply to work properly. Inflammation causes swelling of the blood vessel walls, reducing or even blocking the flow of blood to the tissues and organs (see Figures 2 and 3).

The amount of damage vasculitis causes depends on which part of the body is affected. The larger the affected blood vessels, the more damage there may be. And the more important the affected body tissue, the more serious the damage will be.

The walls of affected blood vessels can swell and bulge (this is called an aneurysm) and may even burst, causing bleeding inside your body. Apart from the damage to the blood vessel itself, this can lead to damage in the tissues or organs that are supplied by the blood vessel.

Vasculitis can appear suddenly in someone who has previously been completely well – doctors call this primary vasculitis. Vasculitis can occur on its own (primary vasculitis) or alongside other conditions (including rheumatoid arthritis, systemic lupus erythematosus (SLE or lupus) or Sjögren’s syndrome) in which case it’s known as secondary vasculitis.

See Arthritis Research UK booklets: *Lupus (SLE)*; *Rheumatoid arthritis*; *Sjögren’s syndrome*.

**Figure 3**

Vasculitis means inflammation of the blood vessels.

The diagram shows a normal artery (top) and one that is inflamed (bottom).
The symptoms of vasculitis vary according to which part of the body is affected.

What are the symptoms of vasculitis?

When any part of your body is inflamed, it swells and is uncomfortable or painful. With many types of vasculitis the swelling is internal and there are no external symptoms visible.

Vasculitis takes different forms according to which blood vessels are affected, and so the symptoms vary from person to person. Many people with vasculitis feel unwell and have fever, sweats, fatigue and weight loss. Sometimes these can be the first symptoms experienced, so it’s important to be seen by your GP. Other symptoms vary according to which part of the body is affected:

- **Skin** – Vasculitis in the skin causes a rash of spots that sometimes burst, leaving open sores (ulcers). When vasculitis only affects the skin, the long-term effects aren’t usually serious, and the symptoms normally clear up once the inflammation has settled.

- **Nose** – Vasculitis here causes crusting inside the nose and nosebleeds. The shape of your nose can change.

- **Fingers and toes** – Some people with vasculitis experience Raynaud’s phenomenon, where the fingers or toes turn white or blue and may tingle or hurt when exposed to cold conditions.

- **Eyes** – Some types of vasculitis can suddenly affect your vision or cause your eye to become red or painful. It’s important to see a doctor quickly if this happens.

- **Nerves** – Inflammation of the nerves can cause tingling (pins and needles) or weakness in the arms and legs.

- **Muscles** – Inflammation here causes muscle aches, and eventually your muscles could become weak.

- **Lungs** – Inflammation of the lungs causes coughing and shortness of breath.

- **Brain** – Occasionally the blood vessels in the brain can be affected, causing problems like strokes.

- **Kidneys** – When vasculitis affects the kidneys there may be problems passing urine or there may be blood in the urine. Vasculitis of the kidneys can be dangerous because the symptoms may not appear until the kidneys have already been damaged. In severe cases treatment on an artificial kidney (dialysis) machine may be necessary.
Headaches, pain in the jaw and problems with the eyes are all possible symptoms of a type of vasculitis called giant cell arteritis (GCA).

What types of vasculitis are there?

Doctors usually define the types of vasculitis according to the size of the blood vessels involved (see Figure 4). The most serious types of vasculitis involve both medium and small arteries.

**Figure 4** Types of vasculitis can be defined according to the size of the blood vessels involved

**Vasculitis in large arteries:**
- includes giant cell arteritis (temporal arteritis) and Takayasu’s arteritis.

**Vasculitis in small arteries:**
- includes Wegener’s granulomatosis, microscopic polyangiitis and Churg-Strauss syndrome
- can also be a result of rheumatic diseases
- can happen with infections including hepatitis and, very occasionally, with different types of cancers.

**Vasculitis in medium-sized arteries:**
- includes polyarteritis nodosa and Kawasaki disease
- medium-sized arteries can also be involved in vasculitis which occurs with rheumatoid arthritis, lupus (SLE) and Sjögren’s syndrome.

**Vasculitis in small vessels (usually capillaries):**
- includes Henoch–Schönlein purpura and cryoglobulin-associated vasculitis
- usually involves the skin and is also sometimes caused by a reaction to certain drugs.

See Arthritis Research UK booklet Raynaud’s phenomenon.
Takayasu’s arteritis
This affects the main artery from the heart and its large branches, usually in younger women. It’s rare in the UK but is more common in the Far East and Africa. The major arteries narrow and this reduces the blood supply to the limbs and other parts of the body. The narrowing develops slowly and the arteries don’t usually block completely, so there isn’t usually a dangerous loss of blood supply to the arms or legs or any major organs.

Giant cell arteritis (temporal arteritis)
This affects the large arteries that supply the head and neck, especially the temporal artery which is found over the temples (see Figure 5). It’s more common in northern Europe and doesn’t normally affect people below the age of 50.

Giant cell arteritis can cause headaches and is often associated with a condition called polymyalgia rheumatica (PMR), which causes inflammation and stiffness in the muscles of the shoulders and hips. Giant cell arteritis occasionally involves the blood supply to the eye, where it can cause blindness. If you develop symptoms in your eyes such as blurring or double vision you should see your doctor straight away as you’ll need to be treated urgently. Other blood vessels such as the major arteries can be involved in giant cell arteritis.

See Arthritis Research UK booklets Giant cell arteritis; Polymyalgia rheumatica (PMR).

Figure 5
Headaches can be caused by giant cell arteritis (temporal arteritis).
Polyarteritis nodosa (PAN)
This condition is potentially very serious but is very rare – only about 1 out of every 2 million people in the UK develop PAN each year. It causes inflammation in the medium-sized arteries, particularly those supplying the gut and kidneys. This may only affect part of the wall of the artery, which becomes weak and may bulge, forming an aneurysm. If it bursts it can cause serious internal bleeding. PAN can also involve the whole wall of the artery at a particular point, which causes a blockage.

Kawasaki disease
Kawasaki disease affects small and medium-sized arteries in children under 5. It’s sometimes called mucocutaneous lymph node syndrome (because it involves the mucous membrane).

Children with Kawasaki disease will feel unwell – they may have a high temperature, swollen glands in the neck (lymphadenopathy), an inflamed area around the eye and the mouth, and a skin rash similar to measles.

This condition is quite rare but can be serious if the arteries supplying the heart are inflamed (coronary arteritis). Up to 60% of patients with Kawasaki disease have coronary arteritis.

Wegener’s granulomatosis
This condition is quite rare and is slightly more common in men than in women. It usually develops with ear, nose and throat problems, including nosebleeds and crusting of the nose, and occasionally coughing up blood (haemoptysis).

These symptoms can appear a year or two before more general vasculitis starts. This general vasculitis usually involves the skin, lungs, eyes and kidneys. The kidney problems can sometimes lead to kidney failure if they’re not recognised early.

Churg–Strauss syndrome
This condition causes asthma to develop in adults, followed by inflammation of the blood vessels caused by swellings called granulomas. There will also usually be a high number of eosinophils (a particular type of white cell) in the blood.

The condition differs from Wegener’s granulomatosis because of the asthma. Unlike Wegener’s granulomatosis, Churg–Strauss syndrome rarely causes damage to your ears and nose.

Churg–Strauss syndrome can also affect the nerves, causing weakness, pins and needles or numbness. There’s also a higher risk of your heart being involved, which can sometimes cause damage to the heart muscle similar to the damage that occurs during a heart attack.
Microscopic polyangiitis
Almost all people with this condition have kidney problems that can lead to raised blood pressure and kidney failure. People may find that they’re tired because of anaemia. Blood tests will show that the kidney is inflamed. Microscopic polyangiitis can also involve the lungs, with bleeding that can cause breathlessness.

Cryoglobulin-associated vasculitis
In this disease, small-vessel vasculitis is associated with cryoglobulins – these are proteins in the blood that stick together in the cold. Having cryoglobulins can reduce the flow of blood or even block the blood vessels, causing damage to the organs or body tissues.

Henoch–Schönlein purpura
Henoch–Schönlein purpura (HSP) affects the small blood vessels (capillaries). It often follows a chest infection and may be an allergic reaction to a virus, food or drugs. It mostly affects children aged 2–10 years, and boys are affected more often than girls. However, adults can also be affected.

The symptoms of HSP include:
• a skin rash often over the buttocks (starts red but develops into a bruised purple colour and appears over several days or even weeks)
• short-lived arthritis, especially of the larger joints
• stomach pain and/or vomiting or passing blood in stools
• passing blood in urine (indicating kidney problems)
• fever, headaches and loss of appetite.

In most cases the condition doesn’t need specific treatment, although relapses are possible for up to a year after the original illness. Kidney problems are quite common, but serious kidney damage is rare. Occasionally other blood vessels are involved, and rarely more serious complications can occur, sometimes affecting the bowels or causing seizures.

Who gets vasculitis?
Vasculitis is rare. In every 100,000 people in the UK, only 5 will develop vasculitis each year – that’s about 3,000 people in all. However, about 22 people per 100,000 will develop GCA*. The different types of vasculitis tend to affect different age groups, for example:
• GCA (temporal arteritis) is much more common in people over 50, and it’s fairly common for it to be associated with a condition called polymyalgia rheumatica (PMR)
• Takayasu’s arteritis tends to affect younger women
• Henoch–Schönlein purpura (HSP) is much more common in children than in adults.

What causes vasculitis?
There’s no single cause of vasculitis, and in most cases the exact cause is unknown. We know that vasculitis isn’t directly inherited, but genetic factors do play a part as several cases can occur in the same family. Genes could make you more likely to develop the condition, in which case it may only take a small trigger (such as an infection or drugs) to start this off.

We also know that some types of vasculitis – for example, those affecting the small blood vessels – can be related to infections, particularly those associated with hepatitis. Some cases of vasculitis follow use of certain drugs, for example propylthiouracil (used to treat thyroid disease) and allopurinol (used to treat gout).

It’s thought that most forms of vasculitis are a type of autoimmune disease. This means that your body’s defence mechanisms aren’t doing their normal job of fighting infections, but instead attack a healthy part of the body, causing inflammation.

See Arthritis Research UK booklet and drug leaflet Gout; Allopurinol.
What is the outlook?
The most severe types of vasculitis can be life-threatening. However, most types respond well to treatment, and for many of them you’re likely to make a full recovery. The outcome depends on the type vasculitis and how it affects you. Overall, the best way to learn more about what might happen to you in the future is to talk to your doctor or another health professional.

How is vasculitis diagnosed?
If you think you may be developing vasculitis you must see your doctor as soon as possible. Infections, drugs and some foods can sometimes cause vasculitis, so your doctor will probably ask about the medications you’ve been taking and your general health during the past few weeks.

What tests are there?
There are many tests that may be done to help diagnose the condition. In this section we’ll look at the most common, but there are others that you may need. Ask your rheumatologist or GP if you’re not sure about what a certain test will involve.

Blood tests may be used to measure inflammation, for example:
- erythrocyte sedimentation rate (ESR)
- C-reactive protein (CRP).

A full blood count can help to establish whether you have anaemia and whether you have normal levels of white blood cells (which fight infections) and platelets (which are involved in clotting).

Blood tests for antineutrophil cytoplasmic antibodies (ANCA) are important in the diagnosis of some types of vasculitis, particularly Wegener’s granulomatosis.

If you have vasculitis along with other conditions, such as rheumatoid arthritis or lupus, then blood tests might be used to assess how active these other diseases are. Blood tests can measure the level of rheumatoid factor in rheumatoid arthritis, or the levels of complement (an enzyme system or group of proteins in the blood) and antibodies in lupus.

Blood tests may be repeated from time to time to check how your condition is responding to treatment.

Other tests may be carried out to see how the affected body organs are working – for example:

- Urine tests – These will show the presence of blood and/or protein, which are often the first signs of an inflamed kidney
  - A urea and electrolytes (U&E) test or a creatinine test may be used to check how your kidneys are working.
  - People with Churg–Strauss syndrome, Wegener’s granulomatosis or microscopic polyangiitis will have regular urine tests for blood and protein.
Treatments are given in stages to keep your vasculitis under control.

- **Liver function tests** may also be carried out.
- **X-rays, CT and MRI scans** can be used to check for chest problems.
- **Echocardiogram and electrocardiogram** – The heart can be assessed by a special ultrasound test known as an echocardiogram and an electrical test, the electrocardiogram or ECG.
- **A biopsy** may be needed to confirm whether the kidneys, the muscles, skin or lungs are affected by vasculitis. A small piece of tissue is removed from the organ in question for examination or testing in a laboratory.
- **An ear, nose and throat (ENT) assessment** may be needed for people with Wegener’s granulomatosis who have symptoms in these parts of the body.
- **An angiogram** is often done where abdominal organs such as the kidney and gut are involved. This involves injecting dye into the arteries so that they show up on an x-ray. They can also be done in Takayasu’s arteritis and giant cell arteritis to see how much the large blood vessels are involved.
What treatments are there for vasculitis?

The treatments used for vasculitis will depend upon which blood vessels and organs are affected, as well as how much body tissue is affected. If the vasculitis only affects the skin, it may be enough to treat any underlying infection or to remove the drug that triggered the vasculitis. However, in most cases, drug treatments will be needed to control the disease and its symptoms and to limit the damage caused by vasculitis.

Drugs
If you have small-vessel vasculitis, then you may only need a small dose of steroids to control it, if treatment is needed at all.

You may be given a combination of drugs, probably over several years. For many types of vasculitis, including those affecting the kidney, lungs or other vital organs (especially if it involves both small and medium-sized blood vessels), your treatment will be given in stages (see Figure 6).

A few people with the most severe types of vasculitis – for example very severe kidney or lung disease – may need a special form of treatment called plasma exchange (also known as blood washing). This means you’re connected to a machine and blood from your body is passed through the machine and returned back to you, after being cleaned of the factors causing the vasculitis.

Figure 6: The stages of treatment for some types of vasculitis

Stage 1: Induction of remission
Aim: to get the disease under control.
An immunosuppressant agent may be used, e.g. cyclophosphamide, that will suppress the immune system, which is attacking the vessels.
Corticosteroids may be used in this stage (tablets or given intravenously).

Stage 2: Maintenance of remission
Aim: when your disease is under control (in remission) the aim is to keep it under control. Milder drugs may be used, e.g. azathioprine

Stage 3: Follow-up and withdrawing therapy
Aim: to keep your vasculitis under control while gradually decreasing the amount of treatment.
Vasculitis varies from one person to the next and from one type of vasculitis to another, so it’s important to follow your doctor’s instructions carefully...

...and to make sure your treatment isn’t interrupted.
If you have vasculitis that mainly affects medium-sized arteries, then other treatments can help, depending on the condition:

- Kawasaki disease can be treated effectively with injections of immunoglobulin (a type of protein).
- Hepatitis-associated polyarteritis nodosa can be treated with antiviral treatment and plasma exchange.

If you have vasculitis affecting the large blood vessels, then you’ll probably be given corticosteroid tablets. These are very effective for giant cell arteritis and Takayasu’s arteritis.

In some types of vasculitis (e.g. Wegener’s granulomatosis) an infection may trigger a relapse. You may therefore be given antibacterial drugs such as co-trimoxazole to protect against this. These drugs can also help to protect against the side-effects of the stronger immunosuppressive drugs.

As with all medications, there may sometimes be side-effects. Cyclophosphamide, for example, can cause bleeding from the bladder, hair thinning and an increased risk of infection. Unfortunately there’s also a significant risk of infertility. Because of these risks, cyclophosphamide will be stopped or exchanged for a different immunosuppressive drug as soon as your vasculitis is controlled. This is usually azathioprine, but methotrexate or mycophenolate might be used instead. Rituximab, a drug given by infusion, can also help to encourage remission in some types of vasculitis.

See Arthritis Research UK drug leaflets Azathioprine; Cyclophosphamide; Methotrexate; Mycophenolate; Rituximab.

Possible side-effects of corticosteroids include weight gain, indigestion, diabetes, thinning of the skin and thinning of the bones (osteoporosis). If high doses of corticosteroids are given, then you’ll also be given drugs like bisphosphonates to help prevent osteoporosis.

See Arthritis Research UK booklet and drug leaflet Osteoporosis; Steroid tablets.

Self-help and daily living
If you do need treatment then it’s very important that you follow your doctor’s instructions carefully. Vasculitis varies from one person to the next and from one type of vasculitis to another. It’s important to speak to your doctor or other healthcare professional about any new symptoms you may have. It’s also important to make sure the treatment isn’t interrupted.
Exercise
Vasculitis can cause tiredness, and it’s important to rest when you need to. However, you should also try to keep muscles and joints healthy by exercising. Start gently and gradually increase the amount of exercise you do. Include some weight-bearing exercise (anything that involves walking or running), and swimming is also recommended. Ask your doctor for advice on how much exercise you should expect to be able to do.

A healthy, balanced diet is important for everyone, but if you’re on steroids it’s particularly important because these can increase your appetite and cause weight gain. Try not to eat too much, and cut down on fatty and sugary foods. Instead, eat more fresh fruit and vegetables and starchy foods like bread, potatoes, rice and pasta. If you’re taking steroids then you’re more at risk of osteoporosis, but having plenty of calcium in your diet is important to help prevent this from developing. Foods that are good sources of calcium include tinned sardines (with bones), skimmed milk, yoghurt and certain vegetables such as broccoli.

Stop smoking
Avoid smoking. It makes the blood vessels become narrower inside and can therefore make vasculitis symptoms worse.

Keep warm
If your fingers or toes turn blue in response to the cold, this may be due to Raynaud’s phenomenon. Wearing warm clothes, including warm socks and gloves, should improve blood circulation to your hands and feet by helping to keep the blood vessels open.

Gentle exercise can help keep muscles and joints healthy.
Research and new developments

In the past, some types of vasculitis were very serious diseases, especially if it affected both small and medium-sized arteries. Treatment over the last 2 decades has completely changed this and most forms of vasculitis can now be controlled and sometimes completely cured. The new problem to be faced is that some treatments have unpleasant side-effects – some are almost as damaging as the disease itself, though in different ways.

Researchers are currently trying to better understand what causes vasculitis. Comparing what happens in different countries may help to find the answer – for example:

- Takayasu’s arteritis is much more common in the Far East and in Africa than in the UK
- giant cell arteritis is extremely rare in India but very common in northern Europe
- Wegener’s granulomatosis is more common in northern Europe than in southern Europe
- microscopic polyangiitis is more common in southern Europe than northern Europe.

There’s a large international group looking into the genetic side of vasculitis, which aims to find out the causes and also possibly investigate new treatments.

Arthritis Research UK funds a range of research into vasculitis, including studies on treatments and on the causes and frequency of vasculitis. Recent examples include clinical trials of higher doses of cyclophosphamide and research into the effects of environmental factors. If differences such as those above can be understood, this will help researchers to develop better treatments.
Glossary

Anaemia – a shortage of haemoglobin (oxygen-carrying pigment) in the blood which makes it more difficult for the blood to carry oxygen around the body. Anaemia can be caused by some rheumatic diseases such as rheumatoid arthritis or lupus, or by a shortage of iron in the diet. It can also be a side-effect of some drugs used to treat arthritis.

Aneurysm – a balloon-shaped sac that bulges out of a blood vessel wall. Some diseases or infections can weaken the vessel wall, and the pressure of blood causes the weakened section to balloon outwards. The bigger the aneurysm gets, the greater the risk of it bursting, which can result in internal bleeding and other complications.

Antibodies – blood proteins that form in response to germs, viruses or any other substances that the body sees as foreign or dangerous. The role of antibodies is to attack these foreign substances and make them harmless.

Antineutrophil cytoplasmic antibodies (ANCA) – a group of antibodies which attach themselves to molecules in white blood cells rather than to foreign substances. They’re commonly found in blood tests for a number of autoimmune disorders such as vasculitis, although they can’t definitely confirm the diagnosis.

Autoimmune disease – a disorder of the body’s defence mechanism (immune system), in which antibodies and other components of the immune system attack the body’s own tissue rather than germs, viruses and other foreign substances.

Bisphosphonates – drugs used to prevent the loss of bone mass and treat bone disorders such as osteoporosis and Paget’s disease. They work by reducing high levels of calcium in the blood and by slowing down bone metabolism.

Complement – an enzyme ‘system’ in the blood. An enzyme is a substance that speeds up a biological reaction. Complement consists of at least 19 separate proteins and plays an important part in the body’s immune system. It allows foreign particles or micro-organisms to be made harmless, but also generates inflammation. Blood tests can show how much of each of the major elements of complement is present.

Computerised tomography (CT) scan – a type of scan that records images of sections or ‘slices’ of the body using x-rays. These images are then transformed by a computer into cross-sectional pictures.

Corticosteroids – drugs that have a very powerful effect on inflammation. They’re often called ‘steroids’ for short. The adrenal glands in the body produce a natural supply but much larger doses are used to treat autoimmune diseases. Prednisolone is the most commonly used corticosteroid.

C-reactive protein (CRP) – a protein found in the blood. The level of C-reactive protein in the blood rises in response to inflammation and a blood test for the protein can therefore be used as a measure of inflammation or disease activity.
Creatinine test – a urine or blood test used to monitor the kidneys. Creatinine is a waste product from your muscles that is processed by your kidneys, so by checking the levels of creatinine a doctor can tell how well your kidneys are working.

Dialysis – a method of separating particles in a liquid by passing them through a membrane. In kidney dialysis the blood is circulated through a special machine that uses this method to remove waste materials or poisons from the blood.

Echocardiogram – a type of scan that uses ultrasound waves to create detailed pictures of the inside of the heart. This test helps show the structure and movement of the heart.

Electrocardiogram (ECG) – a test that records the electrical activity of the heart using a machine called an electrocardiograph. The aim of an ECG is to detect unusual heart rhythms and to identify heart problems.

Eosinophil – a type of white blood cell, which is able to absorb foreign matter. These cells are involved in allergic responses in the body.

Erythrocyte sedimentation rate (ESR) – a test that shows the level of inflammation in the body and can help in the diagnosis of rheumatoid arthritis. Blood is separated in a machine with a rapidly rotating container (a centrifuge), then left to stand in a test tube. The ESR test measures the speed at which the red blood cells (erythrocytes) settle.

Gout – an inflammatory arthritis caused by a reaction to the formation of urate crystals in the joint. Gout comes and goes in severe flare-ups at first, but if not treated it can eventually lead to joint damage. It often affects the big toe.

Hepatitis – inflammation of the liver. This can cause yellowing of the skin (jaundice), fever, abdominal (tummy) pain and an enlarged liver.

Immunoglobulins – a class of blood proteins that are responsible for immunity to specific infections.

Immunosuppressive drugs – drugs that suppress the actions of the immune system. They’re often used in conditions such as rheumatoid arthritis where the immune system attacks the body’s own tissues.

Inflammation – a normal reaction to injury or infection of living tissues. The flow of blood increases, resulting in heat and redness in the affected tissues, and fluid and cells leak into the tissue, causing swelling.

Liver function tests – blood tests used to check the healthy functioning of the liver. They’re frequently performed to monitor drug treatment.

Lupus (systemic lupus erythematosus or SLE) – an autoimmune disease in which the immune system attacks the body’s own tissues. It can affect the skin, hair and joints and may also affect internal organs. It’s often linked to a condition called antiphospholipid syndrome (APS).
Magnetic resonance imaging (MRI) – a type of scan that uses high-frequency radio waves in a strong magnetic field to build up pictures of the inside of the body. It works by detecting water molecules in the body’s tissue that give out a characteristic signal in the magnetic field. An MRI scan can show up soft-tissue structures as well as bones.

Mucous membrane – the type of membrane that lines areas of the body such as the mouth, nasal passages, stomach and gut, vagina, and passages to the lungs.

Osteoporosis – a condition where bones become less dense and more fragile, which means they break or fracture more easily.

Raynaud’s phenomenon – a circulatory problem that causes the blood supply to certain parts of the body to be greatly reduced. It can make the fingers and toes go temporarily cold and numb and they turn white, then blue, then red. Raynaud’s phenomenon can also occur with the condition scleroderma.

Rheumatoid arthritis – an inflammatory disease affecting the joints, particularly the lining of the joint. It most commonly starts in the smaller joints in a symmetrical pattern – that is, for example, in both hands or both wrists at once.

Rheumatoid factor – a blood protein produced by a reaction in the immune system. About 80% of people with rheumatoid arthritis test positive for this protein. However the presence of rheumatoid factor can’t definitely confirm the diagnosis.

Sjögren’s syndrome – an autoimmune disorder that is characterised by dry eyes and/or a dry mouth, aching and fatigue.

Urea and electrolytes (U&E) – a blood test that mainly monitors the kidneys. This is frequently carried out to monitor drug treatment.

Where can I find out more?
If you’ve found this information useful you might be interested in these other titles from our range:

Conditions
• Giant cell arteritis
• Gout
• Lupus (SLE)
• Polymyalgia rheumatica (PMR)
• Raynaud’s phenomenon
• Rheumatoid arthritis
• Sjögren’s syndrome

Self-help and daily living
• Diet and arthritis
• Keep moving

Drug leaflets
• Azathioprine
• Cyclophosphamide
• Drugs and arthritis
• Local steroid injections
• Methotrexate
• Mycophenolate
• Rituximab
• Steroid tablets

You can download all of our booklets and leaflets from our website or order them by contacting:

**Arthritis Research UK**
PO Box 177
Chesterfield
Derbyshire S41 7TQ
Phone: 0300 790 0400
www.arthritisresearchuk.org

**Related organisations**
The following organisations may be able to provide additional advice and information:

**Arthritis Care**
18 Stephenson Way
London NW1 2HD
Phone: 020 7380 6500
Helpline: 0808 800 4050
www.arthritiscare.org.uk

**Churg–Strauss Syndrome Association (USA)**
PO Box 671
Southampton, Ma
USA
www.cssassociation.org

**PMR-GCA UK**
Centre for Disability Studies
Rocheway, Rochford
Essex SS4 1DQ
Phone: 0300 111 5090
www.pmrgcauk.com

**National Kidney Federation**
The Point, Coach Road
Shireoaks, Worksop
Notts S81 8BW
Phone: 01909 544999
www.kidney.org.uk

**Stuart Strange Vasculitis Trust**
West Bank House
Winster, Matlock
Derbyshire DE4 2DQ
Phone: 01629 650549
www.vasculitis-uk.org.uk

**Vasculitis Foundation**
PO Box 28660
Kansas City
MO 64188-8660
USA
www.vasculitisfoundation.org
We’re here to help

Arthritis Research UK is the charity leading the fight against arthritis. We’re the UK’s fourth largest medical research charity and fund scientific and medical research into all types of arthritis and musculoskeletal conditions.

We’re working to take the pain away for sufferers with all forms of arthritis and helping people to remain active. We’ll do this by funding high-quality research, providing information and campaigning.

Everything we do is underpinned by research.

We publish over 60 information booklets which help people affected by arthritis to understand more about the condition, its treatment, therapies and how to help themselves.

We also produce a range of separate leaflets on many of the drugs used for arthritis and related conditions. We recommend that you read the relevant leaflet for more detailed information about your medication.

Please also let us know if you’d like to receive our quarterly magazine, Arthritis Today, which keeps you up to date with current research and education news, highlighting key projects that we’re funding and giving insight into the latest treatment and self-help available.

We often feature case studies and have regular columns for questions and answers, as well as readers’ hints and tips for managing arthritis.

Tell us what you think of our booklet

Please send your views to: feedback@arthritisresearchuk.org or write to us at: Arthritis Research UK, PO Box 177, Chesterfield, Derbyshire S41 7TQ.

A team of people contributed to this booklet. The original text was written by Dr Richard Watts, who has expertise in the subject. It was assessed at draft stage by consultant rheumatologist Prof. Bhaskar Dasgupta and consultant and reader in rheumatology Dr Gabrielle Kingsley. An Arthritis Research UK editor revised the text to make it easy to understand, and a non-medical panel, including interested societies, checked it for understanding. An Arthritis Research UK medical advisor, Dr Ben Thompson, is responsible for the content overall.
Get involved

You can help to take the pain away from millions of people in the UK by:

• Volunteering
• Supporting our campaigns
• Taking part in a fundraising event
• Making a donation
• Asking your company to support us
• Buying gifts from our catalogue

To get more actively involved, please call us 0300 790 0400 or e-mail us at enquiries@arthritisresearchuk.org

Or go to: www.arthritisresearchuk.org

Providing answers today and tomorrow